BILATERAL OPTIC MALIGNANT ASTROCYTOMA IN A 3 YEAR OLD CHILD WITH NFI – CASE PRESENTATION

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We present a rare case of a 3 year old child with a fast-growing bilateral exophtalmus in the context of neurofibromatosis type 1. We analyze the particularities of the clinical and imagistic aspects of the case, and discuss them in the light of pathological diagnosis (malignant astrocytoma) and imagistic follow-up.

Keywords: malignant astrocytoma, neurofibromatosis type 1, optic nerve, optic chiasm

INTRODUCTION

Neurofibromatosis type 1(NF I) is a common autosomal dominant condition that affects about 1 in 5000 people. Optic nerve glioma is one of the landmark lesions characterizing this disease. They usually occur in children during the first decade of life and are seen in 11–30% of patients with NF I. When associated with NF1, the tumors are more often benign, can be multifocal and bilateral, and are usually found within the optic nerves although they can occur anywhere along the optic pathway, all the way to the visual cortex. Although the unilateral occurrence is a finding quite common in this population (up to 30%) bilateral involvement of the optic nerves is rare. Further, the optic gliomas in NF1 are self-limiting lesions with a very slow growth (which stops in adolescence). As a result exophtalmus is a very rare finding and they require surgical intervention in a very small number of cases.

CASE REPORT

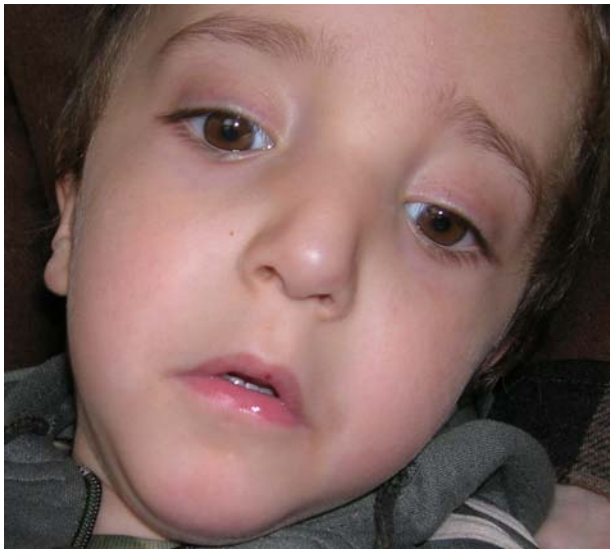
A 3 year old boy was addressed to our department for a fast-growing, grotesque exophtalmus, bilateral but more prominent on the left side. From apparently normal to present status the evolution occurred during a period of roughly 6 months (as documented by family photographs. He lost vision in both eyes 8 months before presentation, after a short period of amblyopia with a rapid course (7 days). The clinical exam revealed café-au-lait spots of various sizes and locations (trunk and limbs) and important of left eye conjunctiva. Both eyes were deviated from the normal position towards inferior left corner. No other clinical sign was apparent.

A brain and orbit MRI exam was ordered with contrast enhancement. Axial T1-weighted magnetic resonance images showed isointense masses developing within both optic nerves, occupying most of the retro-ocular orbit and extended to the optic chiasm. Both lesions enhanced homogeneously after contrast administration. Decision was made to remove both tumors in order to restore facial cosmetics and to prevent further deterioration of the eyes (the left eye was already affected by a severe conjunctivitis). The two gliomas were approached in one surgery through a bifrontal- left pterional approach, orbit ceiling perforation, dural flap and complete tumor removal. The desired cosmetic result was achieved, but third day

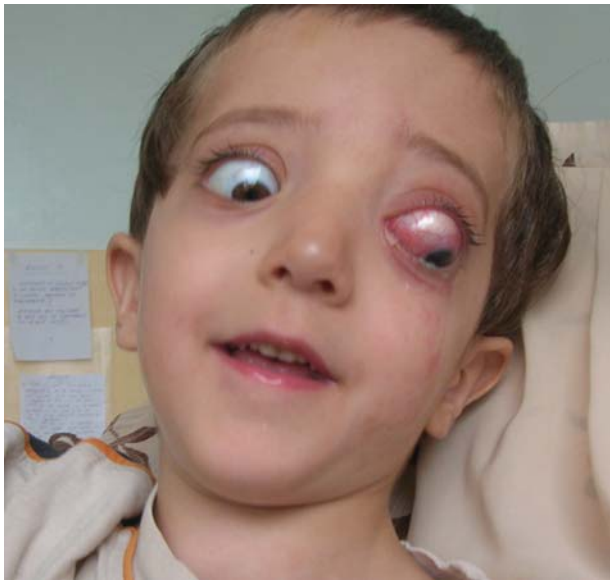
postoperatively the patient developed high fever, vomiting and presented signs of meningeal irritation. Lumbar puncture revealed a high concentration of white blood cells (5100/ml) and immediate antibiotic therapy was started with Vancomycin.

The initial microscopic pathological exam indicated the two tumors as being pilocytic astrocytoma and suggested further investigation through immunohistochemistry. The suggestion was followed and the second exam revealed the tumors to be malignant astrocytomas with a KI67 index of 12%.

At three months postoperatively a MRI follow-up was performed. The new scan revealed a mass occupying lesion within the optic chiasm with the same characteristic as the previous optic nerve tumors. The lesion was, visible on the initial scan, showed an accelerated growth reaching 1,6 cm in diameter. The patient was addressed to gamma-knife center for radiosurgery.



A)



B)

FIG. 1 Clinical aspect in A) November 2008, and B) February 2009

DISCUSSION

Although optic nerve glioma represents a common finding in patients with type 1 neurofibromatosis, either unilateral or bilateral, our case posed a clinical challenge from the start as to the reasons behind a very fast growth rate (from normal to massive exophtalmus in about 6 months). The classic tumor found in NFI is known to be a slow growing tumor that has a self-limiting evolution with variable influence on visual function. Our patient lost vision in both eyes in a very short period (7 days). Furthermore, the MRI aspect of both optic nerve tumors didn't present any particular characteristics to set them apart form the usual optic nerve glioma or to suggest malignancy. Although the initial surgery removed successfully the optic nerve tumors to a good cosmetic result the immunohistochemical diagnostic, the rapid clinical course, and the imagistic documentation of a relapse of the disease at the level of optic chiasm all concur to a reserved vital prognostic. We believe that this case is illustrative for the need to put together all the available data in diagnosing, treating and counseling a case with salient tumors in the context of NFI, never overseeing the most rare possibilities.

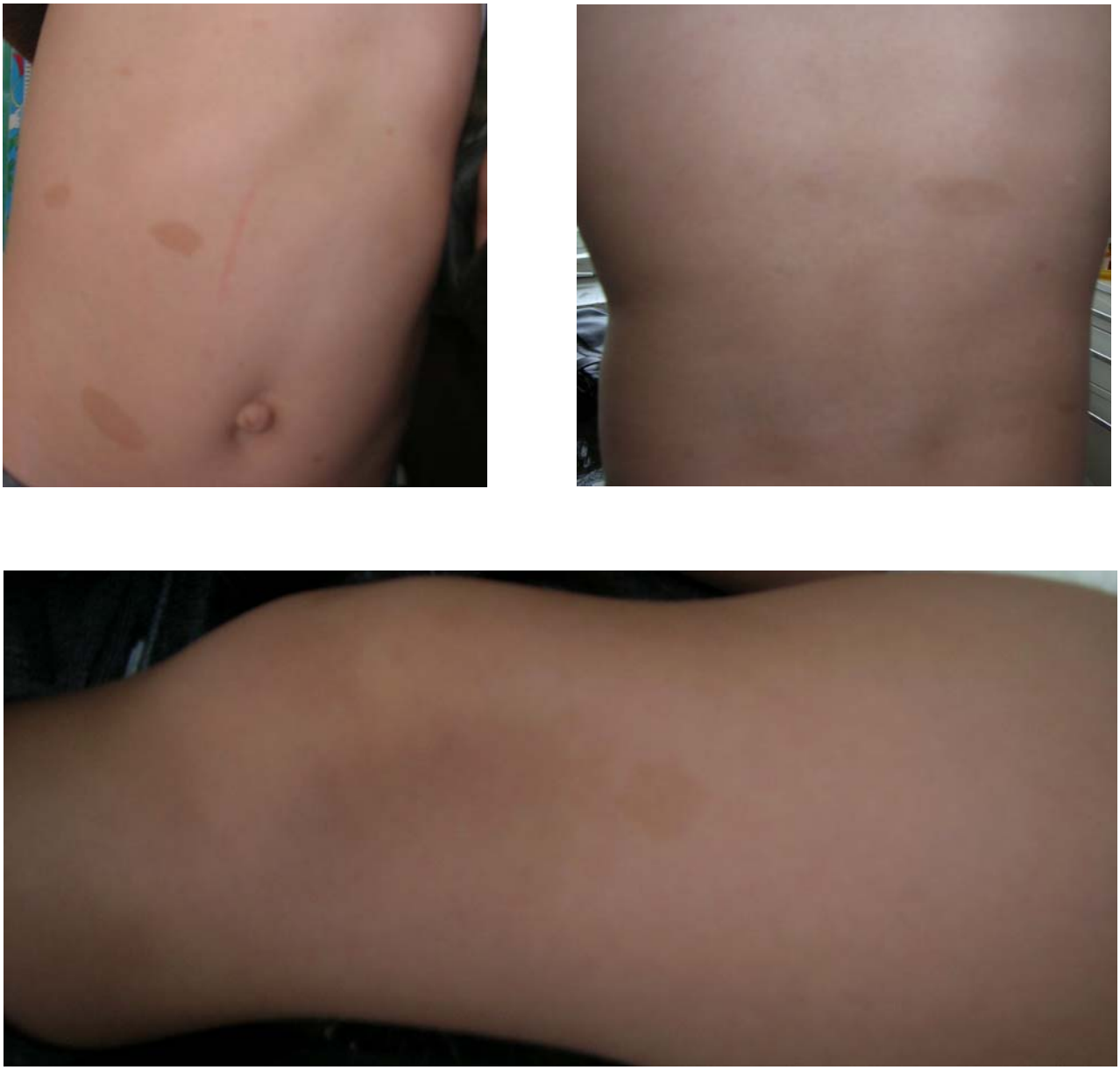
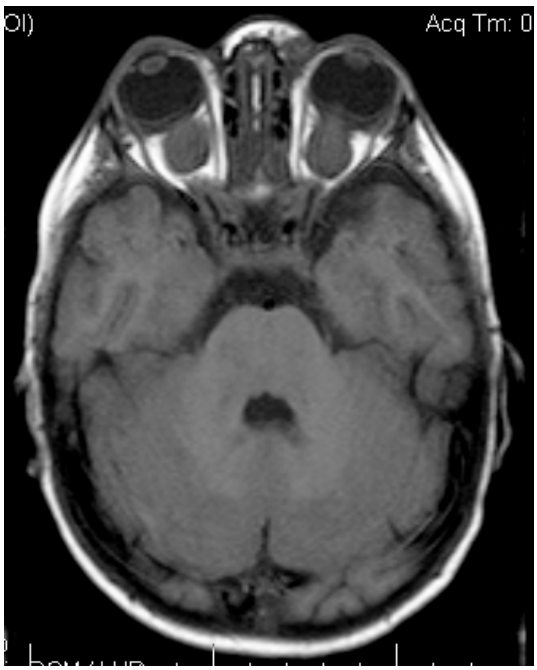


FIG. 2 Café-au-lait spots on runk and limbs



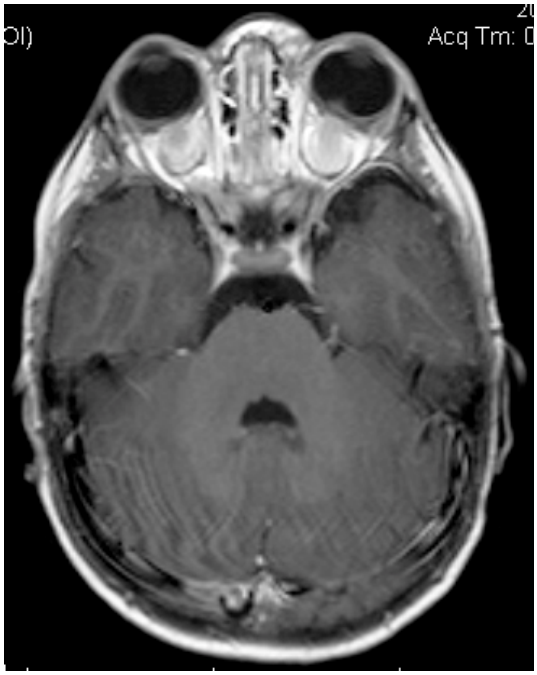
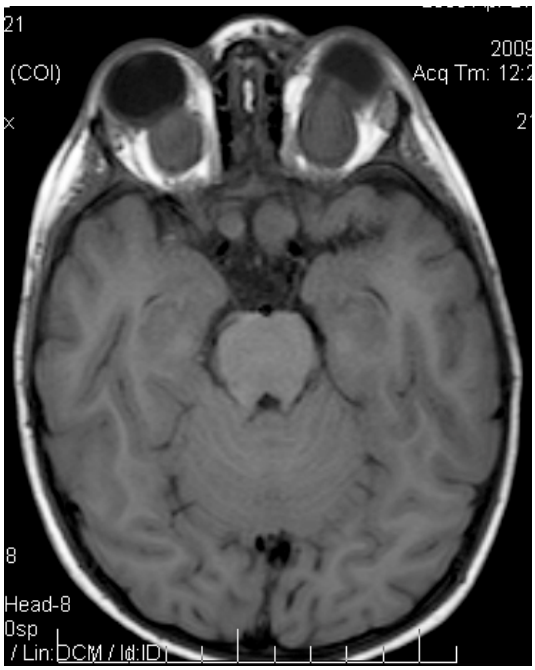


FIG. 3 A)



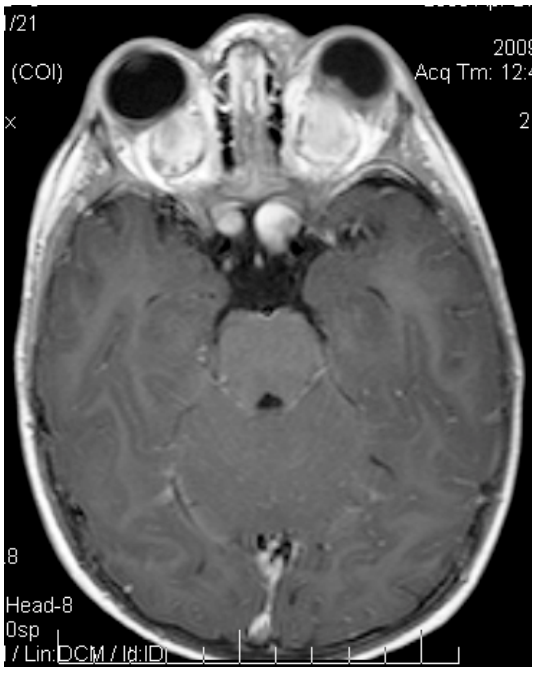


FIG. 3 B)

FIG. 3 Comparative MRI aspects in A) November 2008, and B) February 2009, native and after contrast enhancement on the left and right images respectively

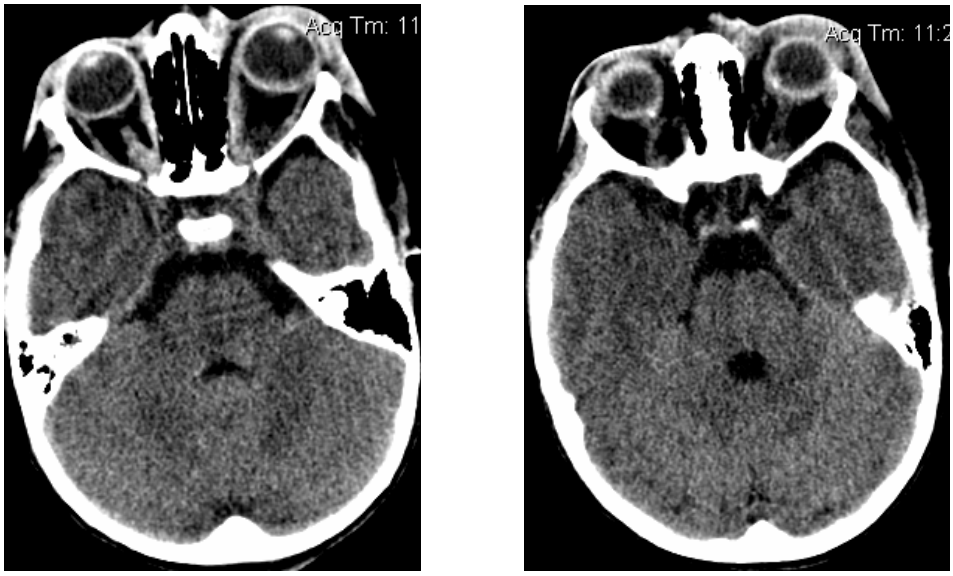


FIG. 4 CT control 1st day postop

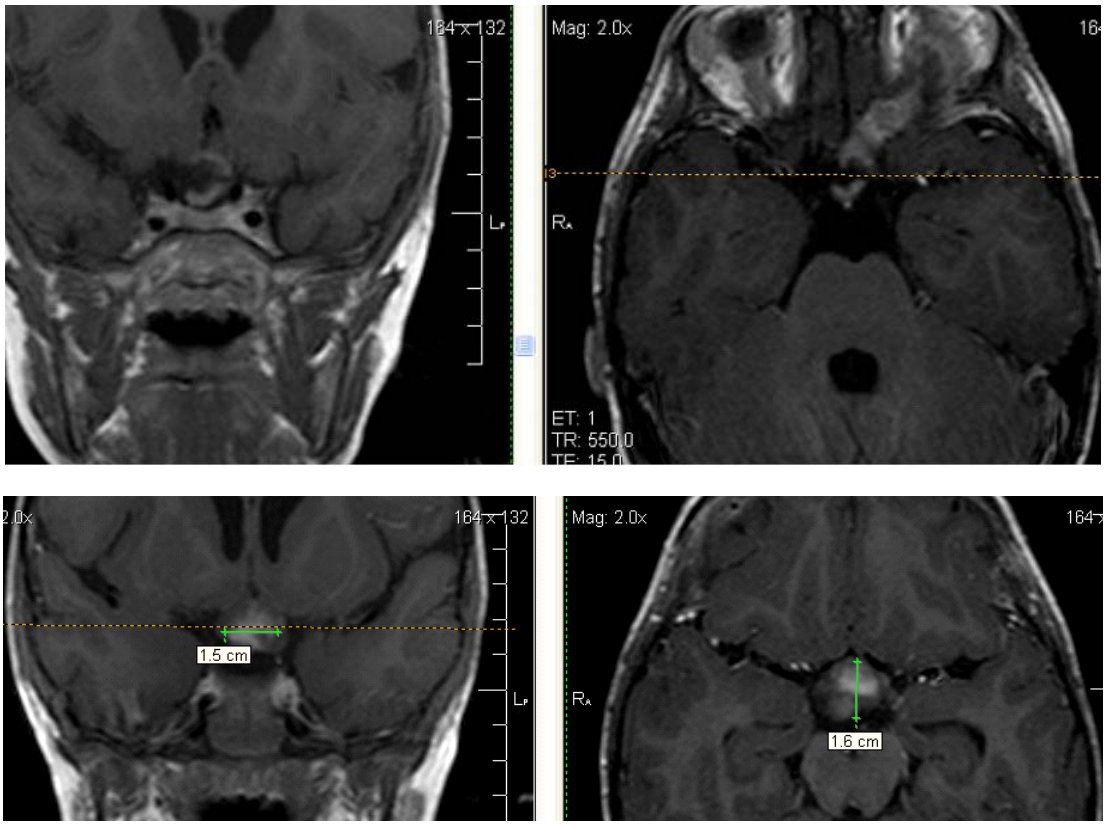


FIG. 5 Mri aspects at 3 months postoperatively